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# Validation of the Italian version of the Cystic Fibrosis Quality of Life Questionnaire (CFQoL), a disease specific measure for adults and adolescents with cystic fibrosis

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**Abstract**

*Background:* Disease specific, health-related quality of life (HRQoL) measurement is important in cystic fibrosis (CF). This work aimed to translate the original English Cystic Fibrosis Quality of Life Questionnaire (CFQoL) into Italian, evaluate the linguistic translation and to psychometrically evaluate the Italian version of the CFQoL.
*Methods:* The linguistic translation followed the international guidelines of forward and backward translation. Psychometric evaluation of the Italian CFQoL involved the assessment of construct validity, internal reliability, concurrent validity, known groups validity and test–retest reliability.
*Results:* The instrument was acceptable to adolescents and adults with CF and demonstrated robust psychometric properties. Principle components analysis indicated that the factorial structure was essentially similar to the original, and the internal reliability of each domain was good (Cronbach alpha coefficients 0.73 to 0.91). Appropriate domains of the CFQoL and SF-36 correlated well indicating good concurrent validity ($r$=0.68–0.80). Consistent with theoretical expectations some domains were able to discriminate between disease severity groups. Test–retest reliability, assessed by intraclass correlation coefficients, was found to be excellent (ICC 0.83 to 0.98).
*Conclusions:* The Italian CFQoL is a valid and reliable measure. Its use in individual patient monitoring and research should complement traditional clinical outcome measures.


*Keywords:* Cystic fibrosis; Health-related quality of life (HRQoL); Linguistic validation; Psychometric validation; Chronic disease

## 1. Introduction

Cystic fibrosis (CF), or mucoviscidosis, is the most common life-shortening autosomal-recessive disease among Caucasians, affecting 1 in about 2500 persons, depending on the country being studied [1], and 1 in 3500–4000 persons in Italy [2]. Once a tragic, poorly understood condition from which people died in early childhood, CF is now a serious, chronic but treatable disorder, affecting many parts of the body such as the lungs, digestive system, sweat glands and reproductive organs. Although advances in the management and care of CF patients have led to the majority surviving into adulthood, cystic fibrosis still involves a complex, time-consuming and, sometimes, difficult range of treatments, limits and rules, and remains a progressive and ultimately fatal multisystem disease with a heavy treatment regimen [3]. This leads to new challenges for people with the disease, their families and the medical teams who care for them. As life expectancy has improved and daily treatments have

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become more complex, understanding the impact of CF on patients' health-related quality of life (HRQoL) has assumed a growing importance [4] and understanding whether this longevity has added "quality years" to the lives of CF patients is a question which needs to be addressed [5].

Health-related quality of life measurement is important in determining the impact CF has on the person's ability to live a fulfilling life [6]. It can complement clinical measures of disease status. Traditional clinical measures such as respiratory function tests and body mass index assessment are important but they do not capture the broader impact of the disease [7]. Generic HRQoL measures have been used because they allow comparison with a healthy population or between different conditions, and because they may detect quality of life issues that were not previously thought to be problematic [8]. However, generic HRQoL measures are not sufficiently specific for a CF population. A few CF specific assessment instruments have been developed or evaluated in different countries in the last 10 years, in an attempt to better understand CF specific issues in clinical practice and research [8–16]. One of these tools is the Cystic Fibrosis Quality of Life Questionnaire (CFQoL) [16], a health-related quality of life instrument for adults and adolescents (age $\geq$14 years) with cystic fibrosis, developed in the UK. The CFQoL is a patient derived measure which includes domains and response scales that have been determined by people with CF and are therefore meaningful to them. The measure includes domains which concern the wider impact on patients' lives (e.g. interpersonal relationships, career issues and future concerns) not found in other CF specific scales.

Cross-cultural validation of an existing valid and reliable HRQoL measure has the advantage of avoiding the time-consuming initial stages of development of a new questionnaire, and allows researchers to choose the measure they are going to validate after considering their own requirements and the psychometric properties of the instrument. This work aimed to 1) translate the original English CFQoL Questionnaire into Italian and evaluate the linguistic translation and 2) to psychometrically evaluate the Italian version of the CFQoL Questionnaire in the Italian CF population.

## 2. Methods

### 2.1. Linguistic validation

A translated questionnaire needs to demonstrate "equivalence" with the original in order to permit a valid comparison of any data derived from them. The linguistic validation of the CFQoL followed the international guidelines suggested by Guillemin et al. [17] and by the MAPI Research Institute [18], and was carried out through the following steps. 1) Two independent forward translations from English into Italian were produced by two professional translators. 2) A first review of these two raw versions was done independently by three medical doctors (a CF Paediatrician, a Pulmonologist and a Generic Medical Doctor) who indicated the clarity of each item, and when provided with different ways of translation indicated the most understandable sentence/word. 3) The CF research team merged the two raw versions to create the first raw Italian version. 4) Two new backward translations from Italian to English were undertaken by two native speaking English persons who had been living in both an English speaking country and Italy long enough to know the languages, the habits and the routine daily life of each culture. 5) The CF research team, with the collaboration of two people from the CF team who developed the original CFQoL compared the backward English version to the original English one. 6) The comparison with the original CFQoL led to the second raw translation into Italian. 8) The Italian CF research team and the English CF research team met a few times to discuss and resolve semantic and conceptual discrepancies and produced the final Italian version of the CFQoL Questionnaire.

Finally, face validity was assessed. Testing the face or content validity of a questionnaire evaluates whether the questionnaire appears to measure what it is meant to, and that it is acceptable to people with CF. Consecutive out-patients attending the CF Centre in Cesena were administered the Italian CFQoL. They were interviewed as to the clarity of each item and concept, the clarity of the response options and the risk of misunderstanding the questions.

### 2.2. Psychometric evaluation

This involved determining the construct validity, internal reliability, concurrent validity, known groups validity and test–retest reliability of the Italian CFQoL. Statistical analyses were undertaken using the Statistical Package for Social Science (SPSS version 12.0).

#### 2.2.1. Participants and procedure

Ethical approval was obtained to conduct the research in all CF Units. All CF patients, aged $\geq$14 years, with a confirmed diagnosis of cystic fibrosis, attending one of the four regional Cystic Fibrosis Centres (Cesena, Parma, Trieste and Grosseto) were considered eligible for the study. Patients were enrolled in the study during a Day-Hospital (DH) routine visit if they were clinically stable, as determined by the CF medical team. They were administered both the Italian CFQoL Questionnaire and the Italian Short-Form Health Survey (SF-36) [19–22]. After 2 weeks, participants were asked to complete the CFQoL again and return it by post.

#### 2.2.2. Measures

The Cystic Fibrosis Quality of Life Questionnaire is a health-related quality of life measure consisting of 52 items across nine domains (physical functioning, social functioning, treatment issues, chest symptoms, emotional functioning, concerns for the future, interpersonal relationships, body image, career concerns). Responses to the questionnaire are

measured by a six point Likert scale. Scores are standardized on a 0 to 100 point scale, where the higher scores represent a better quality of life (scores of <50 represent negative scores, suggesting that the individuals may be experiencing difficulties within that particular domain). In the original English version, the instrument demonstrated a robust factor structure, strong internal consistency, good concurrent validity when compared with the SF-36 health survey, and good discriminatory ability between patients with different levels of disease severity. Test–retest reliability at 10 days was also found to be robust [16]. The CFQoL questionnaire is brief enough to be applied in clinical settings (completion time of about 10 min) and is simple to administer and score.

The SF-36 is a brief, yet comprehensive generic health-related measure consisting of 36 items across eight domains (physical functioning, role-physical, bodily pain, general health, vitality, social functioning, role-emotional, mental health). Scores are standardized on a 0 (worse possible health state) to 100 (best possible health state) point scale, with higher scores indicating a better quality of life. The SF-36 is the most used and established instrument to assess generic health state and has been shown to be reliable and valid [19–22].

#### 2.2.3. Factorial structure and homogeneity

Construct validity evaluates the robustness of the structure of the measure and determines the domains or subscales (e.g. physical function, chest symptoms) of the questionnaire. The factorial structure was tested by principle components analysis (PCA) with eigenvalues >1 on a varimax rotation. Item to domain correlations were calculated to evaluate the extent to which each item was associated with its domain (or with another competing domain). Correlations were regarded as acceptable if they were at least 0.3. The internal reliability of each domain (the extent to which items within a domain are conceptually related) was assessed using Cronbach alpha coefficients. Coefficients show a good internal reliability when the value is 0.7 or above [23].

#### 2.2.4. Concurrent validity

It is typical to asses the concurrent validity of a new instrument by comparing it with an established and validated questionnaire. The medical outcomes survey Short Form-36 (SF-36) was used as a comparator. Correlation coefficients were obtained for the comparable domains on the CFQoL and SF-36 domains (physical functioning, social functioning, mental health/emotional functioning, total scores).

#### 2.2.5. Known groups validity

The measurement of true construct validity is complex as it is an on-going process of assessing whether the instrument performs consistently with theoretical expectations. For example, some HRQoL domains would be expected to discriminate between levels of disease severity. In a complex disease like CF, disease severity is difficult to define but it may be defined by percentage predicted forced expiratory volume in 1 s ($FEV_1$), or age (as increasing age is typically associated with decreased survival). To assess whether the measure could differentiate between patients according to their $FEV_1$, patients were divided into three groups according to the international categorization: mild ($FEV_1 \geq 71\%$), moderate ($FEV_1 = 41$–$70\%$), severe ($FEV_1 \leq 40\%$). For age, patients were also divided into three groups: 14–19 years; 20–29 years and 30 years and over. One-way analyses of variance (ANOVA) were employed to compare the CFQoL domain scores between the three groups.

#### 2.2.6. Test–retest reliability

The test–retest procedure measures the stability of the CFQoL over time. If a repeat administration of the CFQoL is given whilst the person's health status is stable, similar scores should be obtained for each domain at both time points. The time period of 2 weeks was chosen to be long enough not to introduce memory confounds while being short enough to avoid fluctuations in disease status. Alongside the second administration of the CFQoL, patients were sent a brief questionnaire which asked them whether their health had changed since the first completion of the measures. Using a scale of integers from −7 (negative scores denoted worsening health) to +7 (positive scores denoted better health) patients were asked to rate any changes over the 2 weeks [7=changed a very great deal, 6=a great deal, 5=a good deal, 4=moderately, 3=somewhat, 2=a little, 1=almost the same, 0=no change] [24]. Only patients who scored −1, 0 or +1, reporting 'no change' or 'almost the same', and who completed the questionnaires within 14 days were included. Test–retest reliability was assessed by computing intraclass correlation coefficients (ICC).

## 3. Results

### 3.1. Linguistic validation

The acceptability and face validity of the questionnaire was confirmed by positive comments from the CF team and people with CF. Fifteen consecutive patients reported that the items were relevant for people with CF and there were no items, or response options that were difficult to understand. The Italian CFQoL took approximately 10 min to complete. The acceptability of the instruments is also confirmed by the lack of missing data in the psychometric evaluation. Only one participant had one missing response in the whole data set.

### 3.2. Psychometric validation

Between 2003 and 2005, 120 patients were recruited and gave written informed consent and assent. The sample comprised 64 males and 56 females (37 adolescents aged 14 to 19 years and 83 adults), with a mean age of 25.38 (SD=7.64, range=14–46 years). Mean $FEV_1$ and BMI were 68.02% (range=17–124%) and 20.84 (range=13.90–36.00) respectively.

Table 1
Factorial structure and internal reliability of the Italian CFQoL

| | PF | SF | TI | CS | ER | FC | IR | BI | CI |
|---|---|---|---|---|---|---|---|---|---|
| Cronbach alpha coefficients | 0.90 | 0.86 | 0.81 | 0.91 | 0.90 | 0.90 | 0.90 | 0.73 | 0.76 |
| PF1 | **0.66** | 0.43 | 0.02 | 0.13 | −0.05 | 0.18 | 0.12 | −0.05 | 0.09 |
| PF2 | **0.88** | −0.01 | 0.06 | 0.05 | 0.10 | 0 | 0 | 0.27 | 0.08 |
| PF3 | **0.90** | 0.01 | 0.19 | −0.01 | −0.07 | −0.02 | −0.01 | 0.13 | −0.01 |
| PF4 | **0.86** | −0.18 | 0.10 | −0.07 | −0.02 | −0.07 | 0.05 | 0.230 | 0.04 |
| PF5 | **0.82** | 0.06 | 0.10 | 0.01 | −0.23 | −0.20 | 0.13 | −0.03 | 0.09 |
| PF6 | **0.25** | −0.02 | 0.14 | 0.07 | 0.67* | −0.07 | 0.11 | 0.04 | 0 |
| PF7 | **0.50** | 0.17 | −0.16 | 0.09 | 0.29 | −0.01 | 0.09 | −0.04 | 0.03 |
| PF8 | **0.69** | 0.29 | −0.15 | 0.26 | 0.20 | 0.15 | 0.08 | 0.01 | 0.07 |
| PF9 | **0.73** | 0.16 | 0.12 | 0.26 | 0.27 | 0.19 | 0.08 | −0.02 | 0.10 |
| PF10 | **0.73** | 0.19 | 0.16 | 0.30 | 0.23 | 0.16 | 0.12 | 0.08 | 0.09 |
| SF1 | −0.21 | **0.60** | 0.07 | 0.27 | 0.23 | 0.14 | 0.38 | 0.04 | 0.04 |
| SF2 | −0.10 | **0.69** | 0.02 | 0.27 | 0.19 | 0.07 | 0.23 | −0.33 | 0 |
| SF3 | 0.03 | **0.71** | −0.03 | 0.24 | 0.23 | 0.12 | 0.21 | −0.28 | 0.04 |
| SF4 | −0.12 | **0.38** | 0.22 | 0.46* | −0.02 | 0.13 | 0.51* | −0.02 | 0.17 |
| TI1 | 0.22 | 0.14 | **0.75** | 0.29 | 0.12 | 0.19 | 0.05 | 0.06 | 0.11 |
| TI2 | 0.38 | 0 | **0.59** | 0.40 | 0.05 | 0.04 | −0.01 | −0.13 | 0.21 |
| TI3 | 0.54* | −0.15 | **0.36** | 0.47* | 0.03 | 0.07 | −0.12 | 0.21 | 0.13 |
| CS1 | 0.64* | 0.28 | 0.17 | **0.40** | 0.02 | 0.23 | 0.16 | 0.03 | −0.03 |
| CS2 | 0.53 | 0.03 | 0.21 | **0.61** | −0.04 | 0.15 | 0.11 | 0.06 | −0.04 |
| CS3 | 0.31 | 0.07 | 0.15 | **0.77** | −0.13 | 0.089 | 0.21 | −0.023 | −0.05 |
| CS4 | 0.57 | 0.06 | 0.20 | **0.61** | 0.01 | 0.07 | 0.12 | 0.19 | 0.11 |
| ER1 | 0.29 | −0.31 | −0.01 | 0.16 | **0.68** | 0.06 | 0 | 0.04 | 0.32 |
| ER2 | 0.08 | −0.07 | −0.02 | 0.23 | **0.73** | 0.15 | 0.12 | 0.21 | 0.21 |
| ER3 | 0.03 | 0.07 | 0.13 | 0.13 | **0.77** | 0.15 | 0.07 | 0.13 | −0.04 |
| ER4 | 0.23 | 0.13 | 0.12 | 0.23 | **0.68** | 0.04 | 0.25 | −0.03 | 0.26 |
| ER5 | 0.60* | 0.02 | −0.09 | 0.04 | **0.54** | 0.13 | 0.12 | 0.09 | 0.02 |
| ER6 | 0.40 | 0 | 0.01 | 0.18 | **0.51** | 0.10 | 0.35 | 0.14 | 0.01 |
| ER7 | 0.37 | 0.02 | 0.16 | 0.18 | **0.64** | 0.12 | 0.15 | 0.12 | 0.17 |
| ER8 | −0.01 | 0.07 | 0.17 | 0.28 | **0.54** | 0.47 | −0.02 | −0.01 | 0.14 |
| FC1 | −0.02 | −0.03 | 0.01 | 0.26 | −0.02 | **0.77** | 0.14 | 0.04 | 0.04 |
| FC2 | 0.16 | 0.08 | 0.01 | 0.07 | 0.07 | **0.75** | 0.26 | 0.00 | 0.08 |
| FC3 | 0.09 | −0.15 | 0.08 | 0.10 | 0.07 | **0.87** | 0.11 | 0.06 | 0.17 |
| FC4 | 0.12 | 0.05 | 0.06 | 0.22 | −0.02 | **0.72** | 0.08 | 0.15 | 0.26 |
| FC5 | −0.12 | 0.11 | 0 | 0.40 | −0.12 | **0.60** | 0.30 | 0.08 | 0.10 |
| FC6 | 0.01 | 0.09 | 0.05 | 0.47* | −0.04 | **0.44** | 0.36 | 0.17 | 0.22 |
| IR1 | 0.03 | −0.09 | 0.05 | 0.42 | −0.04 | 0.12 | **0.71** | 0.06 | 0.11 |
| IR2 | 0.09 | −0.12 | −0.10 | 0.26 | −0.13 | 0.26 | **0.50** | 0.06 | 0.36 |
| IR3 | 0.18 | −0.15 | −0.04 | 0.19 | 0.042 | 0.13 | **0.70** | 0.25 | 0.28 |
| IR4 | 0.20 | 0.06 | −0.04 | 0.03 | 0.13 | 0.17 | **0.81** | 0.2 | 0.13 |
| IR5 | 0.31 | 0.17 | 0.01 | −0.05 | 0.20 | 0.17 | **0.70** | −0.04 | 0.18 |
| IR6 | −0.03 | 0.26 | 0.23 | 0.35 | 0.10 | 0.24 | **0.51** | 0.14 | 0.30 |
| IR7 | 0.15 | 0.08 | −0.06 | 0.37* | 0.39* | 0.12 | **0.30** | 0.40* | 0.14 |
| IR8 | 0.12 | 0.36* | 0.34* | 0.24 | 0.30 | 0.19 | **0.35** | 0.08 | −0.04 |
| IR9 | 0.22 | 0.17 | −0.04 | 0.12 | 0.26 | 0.27 | **0.33** | 0.14 | 0.18 |
| IR10 | 0 | 0.18 | 0.28 | 0.51* | 0.14 | 0.24 | **0.47** | 0.19 | 0.24 |
| BI1 | 0.13 | −0.10 | 0.10 | 0.23 | 0.16 | 0.24 | 0.01 | **0.55** | −0.02 |
| BI2 | 0.17 | 0.28 | −0.14 | 0.05 | −0.14 | 0.2 | 0.30 | **0.60** | −0.05 |
| BI3 | 0.03 | 0.04 | 0.08 | 0.26 | 0.10 | 0.01 | 0.23 | **0.81** | 0.20 |
| CI1 | 0.09 | 0.10 | 0.10 | 0.09 | −0.02 | 0.25 | 0.30 | −0.02 | **0.78** |
| CI2 | 0.130 | 0.03 | 0.12 | 0.12 | 0.11 | 0.22 | 0.29 | 0.08 | **0.78** |
| CI3 | 0.21 | 0.73* | 0.09 | −0.04 | 0.01 | −0.09 | −0.04 | 0.12 | **0.18** |
| CI4 | 0.05 | 0.19 | 0.05 | 0.32 | −0.03 | 0.11 | 0.24 | 0.15 | **0.66** |

Correlations with own domain are in bold print. *Correlations with unrelated domains that are similar to or greater than intradomain correlations. PF = physical functioning. SF = social functioning. TI = treatment issues. CS = chest symptoms. ER = emotional responses. FC = future concerns. IR = interpersonal relationships. BI = body image. CI = career issues.

*3.2.1. Factorial structure and homogeneity*

The solution that emerged from the PCA highlighted 9 domains and accounted for 72% of the overall variance within the data set. The factorial structure was found to be essentially similar to the original one, even if a few items did not show strong correlation with their factor. The internal reliability was found to be robust, with Cronbach alpha values between 0.73 and 0.91. Item to domain correlations were good and are

Table 2
CFQoL summary data, floor and ceiling effects

| CFQoL domains | Mean (SD; range) | Floor effects (% minimum score of 0) | Ceiling effects (% maximum score of 100) |
|---|---|---|---|
| Physical functioning | 86.13 (16.74; 10–100) | 0.0 | 15.8 |
| Social functioning | 87.17 (18.42; 0–100) | 0.8 | 35.0 |
| Treatment issues | 75.00 (22.31; 0–100) | 2.5 | 15.0 |
| Chest symptoms | 75.33 (23.17; 0–100) | 0.8 | 12.5 |
| Emotional responses | 80.19 (17.57; 15–100) | 0.0 | 5.8 |
| Concerns for the future | 54.81 (27.64; 0–100) | 1.7 | 7.5 |
| Interpersonal relationships | 73.39 (20.12; 16–100) | 0.0 | 5.8 |
| Body image | 77.50 (23.42; 20–100) | 0.0 | 33.3 |
| Career concerns | 75.92 (21.66; 15–100) | 0.0 | 25.0 |

presented in Table 1 together with the Cronbach alpha coefficients. For each domain, CFQoL summary data and floor and ceiling effects are given in Table 2.

*3.2.2. Concurrent validity*

The comparable domains of the CFQoL and the SF-36 correlated well. All the correlations were moderate to strong (physical functioning $r=0.73$, social functioning $r=0.68$, emotional functioning $r=0.70$) and very highly significant (all $p$'s$<0.001$), indicating that the CFQoL has good concurrent validity.

*3.2.3. Known groups validity*

Domain comparisons between the three $FEV_1$ groups demonstrated a significant main effect for physical functioning ($F=4.55$, $p<0.01$), where CF patients with mild disease reported statistically significantly higher scores than patients in both the moderate and severe groups. Mean CFQoL domain scores are presented for the three $FEV_1$% predicted groups in Table 3. Except for treatment issues and body image, domain comparisons between the three age groups indicated that the older adults reported a poorer quality of life in comparison with the younger groups: physical functioning ($F=14.47$, $p<0.001$); social functioning ($F=10.54$, $p<0.001$); chest symptoms ($F=3.27$, $p<0.04$); emotional responses ($F=4.26$, $p<0.02$); future concerns ($F=9.21$, $p<0.001$); interpersonal relationships ($F=5.05$, $p<0.01$); career concerns ($F=7.39$, $p<0.001$). Mean CFQoL domain scores are given for the three age groups in Table 4.

Table 3
Mean (SD) CFQoL scores by $FEV_1$% predicted

| Domains | $FEV_1 \geq 70\%$ ($n=52$) | $FEV_1 = 40$–$69\%$ ($n=46$) | $FEV_1 < 40\%$ ($n=22$) |
|---|---|---|---|
| Physical functioning | 91.32 (8.2) | 83.14 (19.6) | 80.11 (20.5) |
| Social functioning | 90.43 (13.9) | 83.57 (24.2) | 85.83 (12.3) |
| Treatment issues | 76.87 (22.6) | 71.58 (24.0) | 77.40 (19.0) |
| Chest symptoms | 78.94 (21.4) | 71.31 (24.8) | 69.72 (22.8) |
| Emotional responses | 80.95 (18.0) | 78.03 (18.7) | 79.30 (14.0) |
| Concerns for the future | 51.98 (30.0) | 53.49 (25.3) | 52.96 (28.2) |
| Interpersonal relationships | 75.79 (17.9) | 71.14 (19.2) | 68.78 (26.0) |
| Body image | 81.27 (21.7) | 77.77 (22.5) | 74.44 (24.1) |
| Career concerns | 76.28 (21.6) | 74.17 (21.8) | 72.78 (23.7) |

Table 4
Mean (SD) CFQoL scores by age

| Domains | 14–19 years ($n=37$) | 20–29 years ($n=53$) | >29 years ($n=30$) |
|---|---|---|---|
| Physical functioning | 90.91 (9.0) | 90.04 (8.9) | 72.81 (25.8) |
| Social functioning | 93.71 (7.5) | 89.50 (12.3) | 74.44 (29.4) |
| Treatment issues | 80.19 (17.5) | 74.80 (21.1) | 68.89 (28.6) |
| Chest symptoms | 82.00 (17.7) | 73.90 (22.0) | 67.22 (29.3) |
| Emotional responses | 86.93 (13.8) | 76.55 (17.4) | 77.04 (20.4) |
| Concerns for the future | 69.05 (27.08) | 45.33 (22.4) | 48.89 (29.3) |
| Interpersonal relationships | 81.72 (17.7) | 70.96 (18.5) | 67.26 (22.2) |
| Body image | 79.43 (23.0) | 75.87 (23.2) | 82.47 (20.7) |
| Career concerns | 85.43 (18.6) | 73.90 (20.8) | 65.56 (22.5) |

*3.2.4. Test–retest reliability*

Eighty-nine patients completed a repeat CFQoL and reported that their health had remained stable over 2 weeks. Table 5 presents the ICC and 95% confidence interval for each domain. Good test–retest reliability was observed for each domain of the CFQoL.

## 4. Discussion

The aims of this work were to translate the original English CFQoL Questionnaire into Italian, evaluate the linguistic translation and psychometrically evaluate the Italian version of the CFQoL Questionnaire. With a representative sample of the Italian CF population [2], the Italian CFQoL has demonstrated a robust factor structure and internal reliability. The factor structure is basically the same as the UK version, with just a few items showing weaker item to domain correlations. This may be expected given that the CFQoL was developed in the UK. Essentially, the Italian CFQoL has been shown to possess good face, content, construct and concurrent validity. It is internally reliable, reliable over time and able to discriminate

Table 5
Test–retest reliability of the Italian CFQoL

| Domains | Interclass correlation coefficients (95% CI) |
|---|---|
| Physical functioning | 0.97 (0.95, 0.98) |
| Social functioning | 0.83 (0.74, 0.88) |
| Treatment issues | 0.94 (0.91, 0.96) |
| Chest symptoms | 0.93 (0.90, 0.96) |
| Emotional responses | 0.95 (0.93, 0.97) |
| Concerns for the future | 0.96 (0.93, 0.97) |
| Interpersonal relationships | 0.98 (0.96, 0.98) |
| Body image | 0.97 (0.95, 0.98) |
| Career concerns | 0.95 (0.92, 0.97) |

between some known groups based on theoretical expectations. Clinical data are diagnostically essential and crucial for patient monitoring, but it may be argued that clinical information is insufficient for whole patient care and management. The person with CF usually brings with them a much more complex situation in relation to their health cognitions and behaviours. Results from the Italian CFQoL will provide patient reported data on the wider aspect of how CF impacts on a person's life and will complement traditional clinical data.

The measurement of true construct validity is complex as it is an on-going process of assessing whether the instrument performs consistently with theoretical expectations. The CFQoL was able to discriminate between groups based on physical functioning, with those in the least severe group reporting a better HRQoL than those in the moderate and severe groups. Although not statistically significant, there were clinically meaningful differences [25] on several other domains (social functioning, treatment issues, chest symptoms, interpersonal relationships and body image) consistent with the hypothesis that more severe disease, as measured by $FEV_1$, would be associated with poorer reported HRQoL. There are several possible explanations as to why statistical significance was not reached with these domains. Sub-group numbers were small, especially for those with severe disease. Alternatively, $FEV_1$ may no longer be the most appropriate 'global' measure of disease severity. The increase in diabetes, *B. cepacia*, osteoporosis, liver disease and other complications associated with CF longevity may complicate the association between $FEV_1$ and disease severity. The age group data, however, was consistent with theoretical expectation, that age is typically associated with progressive disease. The older group reported the poorest HRQoL for all domains except treatment issues and body image.

The data do not allow for formal, direct comparison with the original UK data due to potentially confounding cultural differences and a different time point in the 'evolution' of CF disease and management. As a disease, CF has changed considerably in the 10 years since the UK instrument was developed. People are surviving longer but there has also been an increase in CF complications (e.g. diabetes, liver disease and osteoporosis). In both the UK and Italian data the mean age (with similar age range) was approximately 25 years. The range of $FEV_1$ predicted values were similar but the mean values were different (UK 55%; IT 68%), indicating a less severe Italian sample. Given the possible confounds, data interpretation is difficult, but it is worth noting that for some CFQoL domains the mean scores were similar (physical, social and emotional function and treatment issues) but for future concerns, career issues, interpersonal relationships, body image and chest symptoms, a better HRQoL was reported in the current Italian data, which may reflect an optimism for a better, longer adult life [26–28].

The Italian CFQoL Questionnaire has been shown to be a valid and reliable measure. The measure is appropriate for several different applications. It may be employed during consultations, clinic review or such as transplant review to guide decision making. As all the items are relevant to CF it can provide a broader assessment of the person's health situation and aid the multidisciplinary CF team in identifying problem areas which may need a specific intervention. The nine domains of the CFQoL are independent scales which, together provide a profile of a person's HRQoL. Therefore, individuals can be assessed on a domain or item basis if there are specific difficulties to address. In many cases, having a measure like the CFQoL, allows the patient to think about his own needs and to communicate to the CF team, not only his concerns about treatments and clinical issues but also his psychosocial concerns. Additionally, as the CFQoL Questionnaire is a *specific* CF health-related measure it may help both the patient and the CF team to face some specific topics.

We must also highlight that having a specific tool to better understand people with CF may be extremely important in the management of treatment regimens. A number of studies have investigated factors affecting adherence to treatment in CF. Early research tended to focus on demographic and clinical variables but recent work has demonstrated that there are multiple factors to determinate adherence, such as personal perception of health status, coping style, worries, social reasons, motivation and side effects of treatments [29–31]. The questionnaire is also suitable for cross-sectional studies where group comparisons are being made. Currently, we do not have data regarding sensitivity to change over time, so it is not possible to estimate clinically important differences. However, the results of the test–retest reliability analyses demonstrate the excellent stability of the instrument and this provides the foundation for the evaluation of responsiveness to change in longitudinal research and clinical practice. Research concerning HRQoL in cystic fibrosis in Italy needs further investigations. The validation of the Italian version of CFQoL represents a first attempt to get closer to this important issue.

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